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A CASE OF JUVENILE TABES IN A FAMILY OF NEURO-SYPHILITICS.

FATHER SYPHILITIC, MOTHER TABETIC, BROTHER PARETIC, SISTER HEMIPLEGIC.

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Cases of juvenile tabes are still very rare. After a most thorough search and painstaking labor, Dr. Otto Marburg,¹ of Vienna, who reported a case of his own, has been able to collect only 34 cases. A critical analysis will show that even this number is entirely too large, as some doubtful cases have found a place in his table. In my opinion, 20 would be an approximately correct number of reported genuine cases.

In the following report I have endeavored to ascertain the facts and faithfully to record them, rather than to quote the already voluminous literature on the subject.

I. *Mother*.—R. C., fifty-seven years old, house-wife, first came under my observation in March, 1904.

Family History.—Her mother died of old age at seventy-five; maternal grandmother lived to one hundred and four; maternal grandfather reached old age. Her father died at eighty, but he came of a short-lived family. There were five brothers, of which number one is probably alive. The others died in infancy or by accidents. There were no sisters. She knows of no form of nervous disease or mental derangement in her entire ancestry.

Health History.—From infancy up to married life she enjoyed good health. At eighteen she became the wife of a healthy man. A son was born in this marriage who is now thirty-eight years old and perfectly healthy. The husband died of some acute disease after only fifteen months of married life. The patient remained a widow for nearly twelve years, during which time she followed the occupation of a canvasser. She was always very active and had no disease during her widowhood. In 1877 she married her first cousin. Her new husband was only twenty-five years old, but he was sick most of the time. He admitted having contracted syphilis at the age of twenty-one. She is positive of having noticed on his body peculiar coppery blotches and that his hair had fallen out. She remembers that he suffered repeatedly from sore throat and hemorrhoids (condylomata?). Headaches were constant and both he and his wife made his dissipated habits responsible for them. He was mentally normal, but drank to excess and died at the age of forty-seven from some chronic pulmonary affection, presumably tuberculosis.

Obstetric Record.—Her first son, W., (juvenile tabes) was born May 21, 1878. Her second son, T., (general paresis) was born October 1, 1880. Her daughter, L., (cerebral lues) was born April 19, 1882. She never had any abortion, nor did she ever miscarry. Shortly after her marriage she developed headaches and rheumatic pains and a little later a peculiar "rash" on the body, which was thought to be urticaria and which persisted throughout her first pregnancy. She never noticed a primary sore, nor did she receive antisyphilitic treatment. Shortly before L. was born she was again covered with a peculiar eruption. The menopause occurred at forty-six and was unaccompanied by unusual nervous disturbances.

Present Trouble.—About twelve years ago she developed certain sensory disturbances. She felt a numbness and tingling of the feet, and a sensation as though she was walking on "stuffed cushions." At about the same time, or shortly thereafter, she began to experience prickling and dull pains in the legs which would come at irregular intervals and sometimes felt like millions of needles. During the last ten years she has had the classical lightning-like pains of tabes which were very severe at first, later moderated a little, and now occur mostly at night. For the past five years similar shooting pains have occurred in the upper extremities. About twelve years ago she lost vision of the right eye. A little later the left eye began to fail and gradually became totally blind. About seven years ago peculiar attacks of abdominal pains made their appearance. She describes the sensation as that of "somebody grinding or twisting her insides out," while there is a burning

sensation in the stomach. Occasionally smothering feelings would accompany these attacks. Since last May she has suffered at irregular intervals from classical gastric crises: spells of vomiting, with intense, agonizing pain in the epigastrium, lasting from twelve to twenty-four hours.

Status Præsens.—In general appearance she is a small, poorly nourished old lady of about sixty-five or seventy years—her real age is only fifty-seven. No signs of lues can be discovered. There is no evidence of peripheral arteriosclerosis, but the pulse is feeble, intermittent and 80 to the minute. The heart does not appear enlarged; the apex beat can scarcely be felt. There are no distinct adventitious sounds, but the second aortic sound is slightly accentuated. No abnormal area of dulness over sternum, no tracheal tugging, no visible pulsations can be observed. The pulse is synchronous on opposite sides and of equal volume. The lungs are emphysematous, but the abdominal viscera appear to be normal. There are no bony or other deformities. There is no loss of muscular power. Special senses, with the exception of sight, are uninvolved. Tremor is absent.

Coordination.—A moderate degree of ataxia of station and gait is present. The patient requires a cane to steady herself in standing, and she cautiously takes small steps in walking. In the upper extremities the ataxia is more pronounced. She cannot approximate the tips of index fingers. In the attempt to touch the nose with the forefinger, the right side proves the more ataxic of the two, but even here the ataxia is not extreme in degree.

Reflexes.—Achilles, patellar, and wrist reflexes are absent. Superficial reflexes are all present. Egger's tuning-fork test (small C., 128 vibrations) demonstrates impaired osteosensibility.

Sensation—Subjective.—A peculiar cushion-like feeling under the feet and paresthesia in the fingers is complained of. *Objective.*—(a) Tactile sensibility is impaired in the upper extremities, and particularly in their ulnar distribution. (b) There is analgesia in the ulnar nerve distribution bilaterally, and also in the peroneal distribution; needle pricks are not perceived as painful impressions. (c) Slight trunk anesthesia is present on the anterior surface of the chest, beginning at the third rib and extending downward four fingers' breadth on both sides. (d) Pressure on the ulnar nerve trunk does not cause pain (Biernacki's sign). There is no persistence of painful impressions; no delayed conduction. The sense of position is greatly impaired on the left side, less so on the right side: she is unable to tell the position of digits and phalanges when they are passively flexed and extended. The stereognos-

tic sense is markedly impaired, tactile and pain sense bilaterally reduced in second and third branches of the trigeminus.

The Eyes.—I am indebted to Dr. Thomas A. Woodruff for his ocular findings in 1900, which were as follows: Atrophy of optic discs. Acuity of vision, right eye: hand movements; left eye 20-100. Field of vision showed marked contraction for white with doubtful color perception. In March, 1904, I found a complete optic atrophy. She could barely tell light from darkness with the left eye; the right was doubtful. There were anisocoria (right pupil smaller than left), and pupillary rigidity to both light and accommodation, but no ocular palsies.

Sphincters.—The bladder function is normal. Since last May she has often been compelled to make hurried calls to the toilet-room, or else she would lose some rectal contents. The patient calls these happenings diarrheal attacks; in my opinion they are signs of sphincter incontinence.

Psychic Symptoms.—Intelligence is good and memory seems to be good, but she states that formerly it was better.

The lightning pains, Westphal and Romberg signs, optic atrophy, gastric crises, the characteristic objective sensory disturbances, paresthesia, the absence of motor involvement, make the diagnosis of tabes a certainty. As in so many women, the syphilitic primary sore has been overlooked, but it is noteworthy that there were no abortions and that no antiluetic treatment was given. It must also be noticed that the ophthalmologist was the first to examine the patient and make a diagnosis of tabes, and that the ataxia has not materially advanced since optic atrophy made its appearance.

On the 27th day of March, 1904, I was hurriedly called to see the patient, whom I found in a wretched condition. She vomited continuously, suffered excruciating pains in the epigastric region, and had a violent diarrhea. These symptoms persisted for about a week with but slight remissions. During all this time she could not take any nourishment, and she emaciated rapidly. This attack came to a sudden stop on April 2, when the vomiting ceased and a complete paraplegia, paralysis of the sphincters, and bed-sores developed. Gradually she became stuporous, talked incoherently at times, and occasionally moaned as if in pain. On April 4, 1904, she was transferred to Wesley Hospital, where her condition remained practically unchanged until April 7, when she became completely comatose and died.

During the last few days of her illness the pulse ranged from 108 to 140, the temperature did not rise above the normal until shortly before her death, when it jumped up to 102.5 degrees F. A chemical and microscopical examination of the urine yielded negative results. The blood, which Dr. R. C.

Whitman kindly examined for me, gave: Erythrocytes, 4,448,000; white blood cells 18,944; hemoglobin, 95%. The increase in the whites was almost entirely limited to the polynuclear neutrophiles.

Post-mortem Report.—About seven hours after Mrs. C.'s death the post-mortem examination was held by Dr. W. S. Griswold in my presence.

We found the body extremely emaciated, so much so that the skin could be lifted up in large folds. Post-mortem rigidity was well marked in the ankles and wrists, not very distinct in neck, elbows and knees. Over each gluteal fold and extending about two inches to either side of anus a large bed-sore was observed. There was post-mortem lividity over the posterior surface of the entire thorax. No signs of violence, nor stigmata of syphilis or tuberculosis could be detected. The brain showed no meningeal hemorrhages nor thickening, neither was it edematous. The convolutions were not flattened and the sulci had normal depth. Upon cutting into the brain substance I found no hemorrhages or softenings. The ventricles were slightly distended, but the ependyma appeared normal. The arteries at the base of the brain, and particularly the carotids, showed a moderate degree of atheroma. When the cord was severed from the medulla there escaped quite a large quantity of cerebrospinal fluid. The cord itself appeared somewhat flattened on its posterior surface; the meninges were not affected. Cutting into the cord one can easily make out a distinct sclerosis of its posterior columns.

Microscopically I found the typical degeneration of advanced tabes, involving almost all exogenous fibers and sparing the endogenous fibers of the posterior columns. The degeneration could be followed up to the gracilis and cuneate nuclei. Serial sections made through the medulla and mid-brain revealed a normal condition by the Weigert-Pal stain. Marchi preparations gave a negative finding, showing the degeneration to have been of long standing.

The other viscera were transmitted to Prof. F. R. Zeit for a macroscopic and microscopic examination, who kindly reports as follows:

(1) Simple hypertrophy of left ventricle, slight chronic fibrous endocarditis of mitral valve. The myocardium shows microscopically some cloudy swelling, fragmentation, nuclei and transverse striæ indistinct; no increase of interstitial tissue.

(2) Extensive atheroma of aorta, but no dilatation.

(3) Chronic diffuse nephritis, interstitial type. The capsule strips with difficulty, leaving a granular surface, on which are seen one or two large depressions. The microscope

reveals in the left kidney distended tubules, which are lined with atrophic epithelium; the interstitial tissue is increased. The capsules of Bowman and the walls of the blood vessels are slightly thickened. Passive hyperemia is present. The right kidney shows similar changes, and, in addition, many small scar-like retractions, the size of a millet seed. Numerous atrophic and obliterated glomeruli and uriniferous tubules are seen.

(4) The spleen was found atrophic; weight only 45 gms.

(5) The liver is smaller than normal, and shows extensive fatty infiltration and some albuminoid degeneration.

(6) Vesicular emphysema of lungs with extreme atrophy of alveolar septa and great distension of alveoli. (This probably accounts for the non-recognition of the heart-hypertrophy and for the existence of indistinct apex-beat.)

(7) Gangrenous vaginitis. (This probably occurred shortly before death, and appears to have originated from the bed-sores.)

(8) Submucous hemorrhage into corpus uteri and constriction of cervical os; atrophic ovaries.

(9) Passive hyperemia of the stomach.

(10) Intestines are slightly congested.

Wishing to discuss the case of *juvenile tabes* separately, I shall first take up the records of the other members of the family.

II. T. C., now deceased. Not having had an opportunity to see him myself, I depend for my information upon his mother and an intimate school friend. T. was the second child, born at full term in normal labor, was well in every way and showed no symptoms of nervous or mental disease until he reached the age of eighteen. He was a pleasing, well-mannered boy, was considered bright and energetic, never dissipated, and his sight was excellent until death. At the age of eighteen a gradual change in his character began: the modest young man became a braggart as regards wealth and attainments. His delusions, however, were not fixed. His former energy dwindled into indecision and lethargy. At about the same time some impediment in his speech was observed; his friend says it was slow, interrupted, and was very much like that of a drunken man. He never had epileptoid or apoplectoid attacks. When his former schoolmate—a very intelligent young man—visited him at the Cook County Hospital for the Insane, the then inmate immediately recognized him. However, in a not lengthy conversation which took place, the patient spoke of having in prospect an excellent position and of various other good opportunities awaiting him. At this same visit the patient forgot his friend's name once

or twice, although he had been an intimate friend for years. During a second visit a few weeks later he noticed that T. wandered considerably in his conversation, was indifferent to domestic happenings, and again had to ask for his friend's name, having called him by name but five minutes previously. Five months after the first visit, that is, shortly before his death, the same friend visited him again. This time he could not recognize him at all; he was confused, seemed in dream-land, and his speech was very imperfect, while his face was in a constant "tremble." After an eighteen months' stay at the asylum the patient died in stupor. At no time was he violent, but a gradual simple dementia seemed to have led to the end.

Even without a personal examination one is justified in making a probable diagnosis of general paresis.

The absence of any focal symptoms, such as hemiplegia, monoplegia, etc., ocular disturbances or other signs directly referring to cerebral lues would probably exclude that disease. The course was too rapid for, and differed from that of, dementia præcox, simple melancholia, or any of the other psychoses. The typical psychical symptoms of simple general paresis were certainly present in abundance. The change in disposition, the loss of memory, the speech difficulty, the flickering of the facial muscles, gradual mental deterioration with confusion and complete destruction of the intellect at the last, all these symptoms make almost a positive diagnosis of dementia paralytica.*

A report from the Dunning Hospital confirms the diagnosis and states that the patient became rather filthy towards the last, and that he died February 5, 1901, after a fifteen months' stay at the hospital.

III. *L. C.*, now nearly twenty-two years old, is the youngest child of Mrs. C.

Health History.—She was born at full term in normal labor about two and a half years after T. When a few weeks old she had "snuffles." At nine months she had a peculiar eruption which involved a portion of the trunk, but principally the junctions of skin and mucous membrane, such as the anal margin and oral ring. From her mother's description she appears to have had moist papules, although they were mistaken for hemorrhoids at that time. At eighteen months a chronic hydrocephalus was observed, which caused no inconvenience. Later she had an asthenic type of pneumonia, from which she recovered perfectly. There never were convulsions during infancy and childhood. Her temporary teeth were not particularly slow in coming, but they fell out like shells soon

* See note at end of paper.

after their appearance. At school she was a good and apt pupil, graduated from grammar school at fourteen, and up to her eighteenth year appeared quite bright; in fact, a little above the average of her associates. During childhood she was well except as noted. Menstruation began at thirteen, and has always been regular and painless.

Present Illness.—During the month of August, 1899, she suffered from severe headaches, which were rather irregular in their appearance and not particularly worse at night. In the wake of these headaches she developed a condition of drowsiness which persisted for nearly twenty-four hours. Her mother advised fresh air, and sent her out. After a time, as she failed to return, the mother became alarmed; looked for her, and found her sitting on the door-step. They entered the house together, and then the mother discovered that her daughter's face was "crooked." She was sent to bed for the night, and next morning it was noticed that the entire right side was paretic with the exception of the upper facial territory. However, improvement occurred so rapidly that on the second or third day she could attend to her duties with but little disability. The headaches, which previously had been so intense, became less so and changed to the type of "dizzy" headache. The next month, in September, she had another attack of right hemiparesis, which was rather evanescent, recovery taking place in one day. In December of the same year she had a third and more severe hemiplegic attack on the same side. It came on rather gradually, several hours elapsing before the paralysis was complete. There was no loss of consciousness, but for about twenty-four hours there was complete loss of speech. Then she began to utter some senseless phrases. This was evidently the usual mixed type of aphasia. She could neither fully understand words spoken to her, nor could she express herself in spoken language. Somewhat later this condition became a paraphasia: she would use words to express herself, but they had not the intended meaning. For an entire year her speech was a mere excuse for language. Even at the present time there is a marked defect in speech: she pauses occasionally and makes repeated attempts at the enunciation of certain words before she succeeds. Improvement of the hemiplegia occurred in the following order: face first, then the arm, and last, the leg.

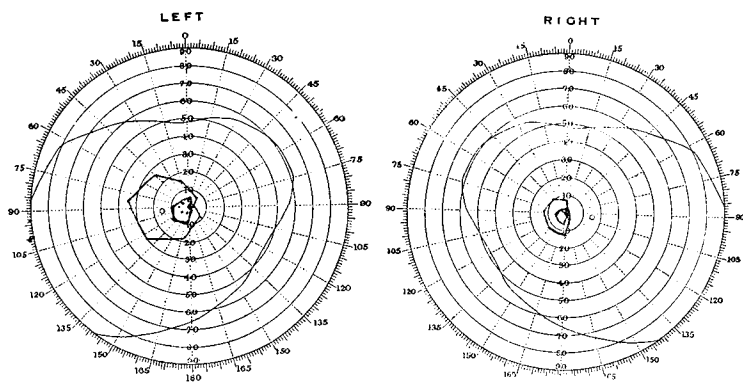
About two and a half years ago she developed a right interstitial keratitis, followed by a similar affection on the left side. She was treated by Dr. T. A. Woodruff and made a good recovery. A little over a year ago she had her first epileptic fit. The attacks at first occurred bi-weekly; later, by appropriate treatment, they were considerably reduced in frequency. The

fits always begin in the paretic right half of the face, then spread to the right hand, arm, and, lastly, to the leg. All this time she is conscious and, so to say, watches her own fit. After the leg has been reached, however, the fit becomes generalized; she loses consciousness and has a typical *grand mal* seizure. Often the attack can be aborted by mechanical means, such as rubbing her cheek, slapping her hands, etc. A great many times these manœuvres are futile and she herself knows that she must have a regular convulsive attack. Immediately preceding the loss of consciousness there is a sensation as though she were going down a precipice—and she falls. The convulsion, which consists of a short period of tonic, followed by a somewhat longer period of clonic spasms, never lasts more than five minutes, and usually leaves her in a dazed condition, which is sometimes followed by sleep of about fifteen minutes' duration. She never bites the tongue, but occasionally froths at the mouth. There is no relaxation of sphincters during the fit. The mother states that the girl's intelligence has somewhat suffered since the last "stroke" of paralysis, and more so since the epilepsy supervened. She is forgetful and somewhat contrary; in fact, her family think she is mentally unbalanced.

Status Præsens.—A well-developed girl, fairly nourished, of average size and rather youthful appearance for her age. There are signs of a past hydrocephalus. Her forehead is rather prominent and makes the face look small by comparison. No radiating white lines are seen about the mouth. The root of the nose is very much flattened and resembles the classical saddle-nose more than a little. A chronic ozena is present and the hard palate presents a distinct aperture. The sternum and both tibiæ are tender on pressure. There are well-marked bony excrescences on the crest of either tibia, giving each a slight convex (saber-shape) curve. Enlarged superficial inguinal glands are distinct. The teeth are poorly formed. The upper central incisors are black, appear crumbled and are notched; the lateral upper incisors are turned out at an angle instead of being parallel, are rather small and far apart. The lower incisors are very thin and present several longitudinal ridges. The tongue does not deviate on protrusion. For the following statement of eye findings I am indebted to Dr. Thos. A. Woodruff. In the year 1900: pupils active. Vision 20-30 and Jaeger I read with difficulty in either eye, disseminated choroiditis and commencing optic atrophy, both sides. Fields of vision for white, green and red are markedly and irregularly contracted, but no central scotomata are present, and no inversion of the color fields. His findings at present (March, 1904), are: with the exception of a number of floating vitreous opacities in both eyes and some haziness of the corneæ, the media are

clear. In the fundus of either eye are seen patches of choroid atrophy, pigmented and optic nerve atrophy. With a high plus lens the Hirschberg vessels in the cornea are easily seen. The pupils are equal, 5 m.m. in diameter, and react to light and accommodation. The fields of vision differ, are very much contracted, more so on the right, as can be seen by accompanying charts. There are no ocular palsies.

Motor Apparatus.—The right upper facial territory is uninvolvement, the right middle and lower facial areas show a flattening out of the lines of expression, and the right angle of the mouth droops. This hemiparesis is made more evident when she is asked to whistle, uncover her teeth, etc. The right upper extremity shows marked weakness as compared to the left. The shoulder muscles are weak, so that she cannot raise her hand above her head. There are no contractures and no atrophies,



but there is some rigidity on passive motion. There are no athetoid or choreic movements of the fingers, but associated movements are very distinct whenever the left hand executes movements. The gait is of the typical hemi-paretic-spastic variety. The patient swings her leg in an arc of a circle, foot in equino-varus position. There is no shortening or atrophy of the paretic limb.

Reflexes.—The wrist, triceps, knee and Achilles reflexes are bilaterally exaggerated; the right more so than the left; no ankle clonus. Neither the Babinski nor Oppenheim sign can be obtained. Heart and lungs are normal. There are no visceral disturbances of any kind.

Sensation.—On the paretic side slight tactile anesthesia and some hypalgesia can be demonstrated.

Snuffles, suspicious eruptions, chronic hydrocephalus, condylomata, chronic oozena, destruction of hard palate, osteophytes on long bones, disseminated choroiditis, deformed

teeth, interstitial keratitis,—point indubitably to lues hereditaria.

The hemiplegia was probably caused by thrombosis with softening and later cicatrization. The gradual onset, the recurrent incomplete attacks, absence of unconsciousness, and the aphasia, point to arterial thrombosis, while the Jacksonian fits would indicate cortical irritation, and in this instance point to the existence of scar tissue in the Rolandic area. The age of the patient (seventeen years), the lues hereditaria, and gradual and repeated development of hemiplegic attacks, and the further course of the disease, make the diagnosis of brain syphilis certain.

IV. *W. E. C.*—Age twenty-five; single; was a telegraph operator until about four years ago. At present he is a medicine and newspaper vender.

Health History.—He was born at full term in normal labor. When about two to three months old is said to have had moist papules around anal margin and rhagades about oral ring. At the age of four months he had a generalized eruption. He had measles at about six years, and typhoid fever eight years ago. At fifteen he had what he calls "shingles," a vesicular eruption. When about five years old and up to his tenth year he suffered from enuresis nocturna and diurna. After this there developed difficulty in emptying the urinary bladder, slight at first, but at the present time more aggravated.

Habits.—For two years, up to quite recently, he was a heavy drinker. For the past four months he has been a total abstainer from alcoholics.

He has never sustained any injuries.

Present Illness.—In 1896 he accidentally discovered that his right eye was totally blind. Shortly thereafter vision began to fail in the left eye and for one year and a half he has been completely amaurotic. About five years ago he developed characteristic lightning-like pains in the lower extremities. They were very severe at first, and each attack would last from two to three days with slight remissions. At the present time he is only infrequently annoyed by these pains and they have lost their former severity. A peculiar numbness and tingling in the lower extremities appeared soon after the shooting pains had begun. Added to this there was a feeling as though he walked on rubber. Quite recently he experienced pains which begin in the hypochondriac region and extend almost circularly to the spine, and which give him a sensation of a band tightening about his body.

Status Præsens.—General appearance rather youthful, beardless and feminine. No asymmetry of face and skull. Radiating from each corner of his mouth are the silvery white lines

typical of inherited syphilis. The teeth are well-formed and there is an abundance of pubic hair—but the testicles are infantile in size. Heart and lungs are normal, the pulse is regular. The abdominal viscera are normal. Bony deformities, and peripheral arteriosclerosis are absent, the superficial inguinal glands are slightly enlarged, but no epitrochlear or cervical glands can be felt. There are no visible scars on the trunk, but at the anal margin are distinct radiating lines, probably scars of former condylomata. There is a perforation of the nasal septum.

Eyes.—Dr. Woodruff, who treated the entire family for eye trouble in 1900, has kindly communicated to me the result of his findings at that time. He found complete optic atrophy in both eyes. Vision of the right eye, hand movements; of the left, 5-200. Fields of vision showed marked contraction for white, with doubtful color perception.

At the present time there is a slight drooping of the left upper eyelid and paresis of the left internal rectus; the eye does not rotate beyond the middle line when the patient looks to the right. When this oculomotor paresis first occurred is unknown, but evidently after he became completely blind, or diplopia would have been noticed. Both pupils are widely dilated, the left a trifle more so than the right; neither responds to light nor to accommodation. The blindness is so complete that not even light can be distinguished from darkness. Both optic nerves present a snowy white appearance with distinct definition, and almost total disappearance of the vessels.

With the exception of the above-mentioned partial paresis of the motor oculi, no muscular weakness can be detected; no tremor in fingers and hands.

Sensation.—Slight paresthesia, lightning pains and girdle sensation are complained of. *Objective.*—Tactile, pain and temperature senses are intact, except that in a few isolated spots on the anterior surface of the legs a pin prick is first noticed as a touch and a little later, as an intense and prolonged pain. Firm pressure of the right testicle causes a sickening pain; pressure of the left is first felt as touch and a little later he cries out from pain. The sense of position is unimpaired in the hands; in the toes of the right foot flexion was mistaken for extension; and he was entirely unable to indicate the position of his left toes. Osteosensibility is greatly reduced according to Egger's tuning-fork test (small C, 128).

Coordination.—Ataxia of station and of motion, already noticed by the ophthalmologist in 1899, is well marked. Though not extreme it can be readily elicited by the heel-to-knee test. In the upper extremities the ataxia is less marked on the right than on the left side.

Under this heading I might also mention a peculiar ataxia of the tongue and facial muscles. When the patient protrudes the tongue there is a constant forward and backward swaying of the tongue, resembling somewhat the lateral swing of a pendulum. It is coarse and dissimilar to any tremors that I have seen.

The facial muscles show a peculiar quiver when in action. Oppenheim mentions the existence of ataxia of facial and tongue muscles in tabes as rare.

Reflexes.—Abdominal and plantar reflexes are normal; the cremaster cannot be obtained on either side. McCarthy's supra-orbital reflex is present. Deep reflexes are absent.

Sphincters.—While the patient suffered from incontinence of urine up to his tenth year, he can now hold his urine very well, in fact, too well. He has great difficulty in emptying his bladder. Constipation is obstinate.

Psychic Functions.—The intellect is not impaired. He is good in arithmetic, reasons well, and attends to his business without mistakes. His usual cheerful disposition has remained unchanged. Although he finds fault with his memory of late, I have been unable to discern any kind of psychic deterioration. About four months ago a peculiar speech disorder developed, which has the attributes of both the paretic and the scanning speech, and which is very difficult to describe. When asked to repeat the usual paradigmata, "Peter Piper," etc., "Round a rugged rock," etc., he shows some degree of syllable slurring. But in conversation he seems rather to scan his words and leaves out no syllable. In the absence of any, even the slightest, psychic alterations, I am led to attribute this speech disturbance to the lingual and facial ataxia mentioned above.

On April 8, 1904, the patient was suddenly taken ill with symptoms of acute cold. Two days later he developed a right lobar pneumonia and was transferred to Wesley Hospital. During his entire illness the temperature did not rise above 102.4 degrees F., but he was delirious most of the time and had to be kept in restraint. There was an uneventful recovery and the patient left the hospital April 24. Upon my visit to his house one week later I found him in excellent spirits. He said his nerves were steady and he felt strong and well. In his demeanor he was rather quiet and so were his facial muscles. The twitching previously observed was reduced to a minimum and even the left-sided ptosis previously noted was ill-defined. The tongue was less tremulous than it had been before his acute sickness, and his speech was almost normal. An improvement in the nervous symptoms of patients affected with chronic nervous disease has often been observed after the subsidence of a complicating acute infectious disease.

Simple enumeration of the symptoms leaves no doubt of the diagnosis of juvenile tabes, to wit: loss of deep reflexes, ataxia, rigid and unequal pupils, primary progressive optic atrophy, ptosis, lightning pains, bladder disturbances, girdle sensation, paresthesia. One must bear in mind, however, the two disorders for which juvenile tabes has so often been mistaken: Friedreich's ataxia and cerebrospinal lues.

Friedreich's ataxia, according to Charcot's² beautiful lecture, may be clinically thought of as a combination of tabes and multiple sclerosis. In this disease there is the hereditary and family element, the ataxia is cerebellar in type and occurs both at rest and in motion; there is a chorea-like oscillation of head and trunk; upper extremities are early involved; kneejerks are lost late; bladder disturbance is rare. There are no lancinating pains; sensory disturbances, if present at all, are slight; there is scanning speech, nystagmus, scoliosis, club-foot, paresis in peroneal region, vertigo and disturbance of intelligence; no amaurosis, optic atrophy, nor Argyll-Robertson pupil. It will be observed that neither this description nor that of an author such as Schmid,³ and others, show any similarities with my case.

As for cerebrospinal lues, it is certainly possible for that disease to assume the mask of tabes until discovered on the post-mortem table; or syphilis and tabes may both be present, as in the observations of Sidney Kuh,⁴ who found degeneration of the posterior columns and, in addition, a distinct cerebrospinal meningitis. Martin Brasch⁵ has described the anatomical findings of syphilis of the central nervous system in a patient who, during life, presented a typical picture of taboparesis. Minor,⁶ Dinkler,⁷ Sachs,⁸ Nonne,^{9, 10} have reported similar cases. H. Schmaus¹¹ even speaks of the difficulty in the correct anatomic interpretation of sclerosis of the posterior columns where luetic spinal meningitis is also present. In the present state of our knowledge, if the lesions of spinal syphilis exactly coincide in location with those of tabes there are no positive differential signs. One might emphasize the importance of searching carefully for evidences of focal disease, such as vertigo, spasm, localized paresis, symptoms indi-

cative of gross brain disease, optic neuritis; that primary progressive optic atrophy is as frequent in juvenile tabes as it is rare in cerebrospinal lues; the history will, in doubtful cases, also give additional help.

Another objection might be urged against my diagnosis of juvenile tabes on account of the speech difficulty and the tremulousness of tongue found in the patient. The point might be made that this is general paresis with absent reflexes, and ataxia. Against this is the fact that there are absolutely no psychic defects and that the patient is capable of managing his business without a flaw. It may be argued that this is tabes complicated by dementia paralytica. There can be no certainty on that point. Eulenburg¹² states: "A facial paralysis in tabes is rather rare, is incomplete and transient. Still rarer is a tongue paresis with disturbances of speech. In these cases the paresis is central." And Charcot² says: "In genuine tabes there is never any speech difficulty. Of course speech disturbances might also be found in genuine tabes, but in those cases there is added to the tabes a general paresis." According to the latter author my case with the imperfect speech and somewhat tremulous tongue and facial muscles would probably be tabo-paresis. On the other hand, Oppenheim¹³ says: "Ataxia but rarely attacks the facial and tongue muscles and leads to a peculiar speech disturbance depending upon excessive movements of lip, tongue and jaw muscles." This must be one of those rare cases. Binswanger,¹⁴ in speaking of tabo-paresis says: "For years there may be only tabes; then develop the ethical defects, the change in the affections, etc., while intelligence and memory may for a long time remain intact." No ethical defects are found in my case.

To sum up, we have an uncomplicated case of juvenile tabes in which syphilis is next to being proven in both ancestors and in all their offspring. The patient himself has the marks of syphilis on his person, however mild the disease may have been. Brasch¹⁵ may be correct in saying, that it almost appears as though those syphilitics who are to develop tabes are by that reason immune from frequent relapses of genuine syphilis.

Referring once more to the frequency of juvenile tabes, it

may be recalled that in 1892 Hildebrandt¹⁶ critically examined all the cases reported up to that time and rejected all but 10 as belonging to Friedreich's disease. S. Kalischer,¹⁷ still further reduces that number. Dydynski¹⁸ reports a case of his own and includes in his report the three cases of B. Remak,¹⁹ one of Strümpell,²⁰ one of Mendel,²¹ one of Bloch.²² All others he counts among Friedreich's disease. And now comes Brasch¹⁵ in 1901, adds a case of his own and after criticizing Kalischer for striking out B. Remak's 3 cases, counts up 8 genuine cases. V. Halban²³ reports 3 cases that appear to have been genuine, and later another case²⁴ which can withstand criticism. Bloch's²² first case is doubtful. In the discussion which ensued after presentation of his case in the Berliner Gesellschaft für Psychiatrie und Nervenkrankheiten in 1896, the late Professor Jolly called attention to the psychic disturbances and the convulsions and referred these to the gross brain lesions of cerebrospinal lues. Professor Oppenheim on the same occasion emphasized the fact that the symptom-complex of tabes in childhood is often due to syphilitic disease of the nervous system; he also doubted the diagnosis. Bloch's²⁵ second case, reported in 1902, seems to be a genuine case, but lacks a history of syphilis. Otto Marburg,¹ describes in detail a case of infantile tabes, but his case is not quite clear. However, he has constructed an elaborate table covering the entire bibliography on the subject. He finds 34 cases in the entire literature and includes in his table some of the doubtful cases, but omits all cases after the age of twenty-one. Dr. R. T. Williamson, of Manchester, reports three genuine cases of juvenile tabes in the June, 1904, number of the *Review of Neurology and Psychiatry*. The words infantile and juvenile, as referred to tabes, are used indiscriminately by most writers, and they all seem to agree that juvenile does not differ from adult tabes in its symptomatology.

Dydynski,¹⁸ however, sums up the peculiarities of juvenile tabes as contrasted with ordinary tabes in the following: (1) Vesical disturbances, and (2) optic atrophy appear early; (3) ataxia is either absent or late in appearing.

From a perusal of the literature I am led to agree with the last-named author that optic atrophy, vesical trouble, sensory disturbances, are among the early symptoms of juvenile

tabes, and that syphilis has invariably been present in the antecedents of all patients. I cannot see how the opponents of the syphilis-tabes theory can ignore the latter fact. It was in order to silence the new opposition emanating from a Berlin clinic that caused Professor Erb²⁶ to come forth with a new series of statistics. He again figures out 90% of syphilis in the etiology of adult tabes and points to juvenile tabes as unqualifiedly caused by syphilis.

One of the most recent writers on the pathology of tabes, Homén,²⁷ accepts the post-syphilitic view of tabes and the theory that the metabolic products resulting from the action of the syphilitic toxins upon the tissues produce the degenerative nerve disorders, tabes and general paresis. The best support for this view can be found in the cases of juvenile tabes where none of the other contributing factors, such as excessive alcoholic and sexual excesses, come into play. The children with congenital or early acquired syphilis who have escaped destruction or severe mutilation have probably harbored an attenuated virus, the chemical products of which were not sufficiently violent to cause inflammations, but were still capable of producing degeneration of nervous tissue, of which tabes is an example.

As regards the oft-debated question, which is the more important, the soil or the morbid agent, in determining the character of the particular nervous disease to be produced, opinions are still divided. While some consider the neuropathic constitution the all-important factor, others lay more stress upon the peculiar quality of the virus as productive of certain nervous diseases. For instance, Babinski,²⁸ in discussing cases of conjugal tabes reported by him and others, says:—"These experiences establish the existence of a special form of syphilis affecting preferably the nervous system, irrespective of the soil. There must, therefore, be a variety of syphilis with a peculiar predilection for the nervous system, and persons contracting syphilis from such source are liable to have nervous syphilis. In the case of syphilis of the conjugal variety—if the syphilis be of the nerve-attacking species—the manifestations will occur in the nervous system, but chance alone will determine whether they will appear as tabes, general paresis,

or other gross brain lesion." And again, A. Souques,²⁹ after having presented before the Neurological Society of Paris a couple affected with conjugal tabes, takes occasion to reflect upon the etiology as follows: "Is it only coincidence when both parties become affected in the same way and is the syphilitic infection caused the same as in single persons, or is there a certain form of syphilis that always selects a certain tissue in the body? It is certain that not all forms of syphilis produce the same effects, for tabes is still rare among syphilitics. It must be admitted that only certain kinds of syphilis seem to enjoy that privilege on account of their great virulence upon nervous tissue." He accepts the hypothesis of MM. Morel-Lavallée and P. Marie, that there is a specific virus of nervous syphilis. To further support his contention he refers to the work of MM. Besançon and Labbé, who proved experimentally that ordinary pyogenic staphylococci taken from a suppurating joint and injected into other animals, either intravenously or subcutaneously, show a tendency to localize themselves around joints. He thinks their previous sojourn in a joint has modified their virulence and communicated to them elective properties. It is possible that the virus of syphilis may in a similar manner acquire a particular preference for the nervous system; and in no series of cases can this be better illustrated than in conjugal tabes where the syphilitic infection is most usually acquired from a single source. Cases of nervous syphilis have been published that were derived from a single source and resulted in the production of identical symptoms. He further refers to Morel-Lavallée who cites 5 cases of general paresis which had their origin in a single source of infection. The following report of Marie and Bernard³⁰ supports the same view. They relate the case of two friends from the country who came to Paris together. Shortly thereafter they contracted syphilis in the same way and from the same woman. In 1890 one had the first symptoms of tabes, the disease showing itself first by ocular disturbances. One year later the other showed symptoms of the same disease which manifested itself in the same manner. Two years later both were suffering from typical tabes dorsalis with the lightning-pains and incoordination well marked.

Max Nonne³¹ relates two similar experiences where individuals became infected with syphilis by one and the same source and later developed either syphilis or parasyphilis of the nervous system. In one case, of the three individuals who together worshipped at the shrine of Venus one became tabetic, the others developed general paresis. In his second observation, of two merchants who exposed themselves to the same source, one developed a severe cerebral lues, the other a syphilitic spinal paraplegia with pupillary rigidity.

Coming back to our case of juvenile tabes and the other nervous disorders in that family, we might ask: which was the more important etiologic factor, the neuropathic family tendency or the specific syphilitic nerve-virus? It would be difficult to give a correct answer. If we are to assume that there is an inherent tendency to nervous disease in that family, which tendency was made still more powerful by the fact that the parents were first cousins, we should probably feel satisfied, were it not for the fact that no neuropathic history can be found in the family record. Might we not consider our case as an additional proof supporting the theory of the French authors that there is a specific nerve-attacking syphilitic poison? The father, being the *fons et origo* of this family's difficulties, died before the so-called syphilitic nerve-virus had created disturbances in his own body, but not before he had transmitted the disease to his wife and children. And now began a process of selection of certain members for certain of the organic syphilitic nervous diseases, which may not altogether have been determined by chance. The mother, for instance, having had a very mild form of lues, but having undergone the stresses of a sharp struggle for existence, developed tabes dorsalis. The first child, a male, developed mild lues and during the stress of puberty, tabes. The second son, the most intellectual of all the children, died of general paresis, a disease that selects with preference a highly developed brain. It is possibly for this reason that savages and semi-civilized peoples have a great deal of syphilis and little or no general paresis. During pregnancy with her last child the mother suffered from a recrudescence of syphilis, as may be recalled from the history, and a girl was born with the stigmata of a rather severe type of syph-

ilis. The brunt of the attack was spent upon her arterial system, causing grave cerebral disturbances—but this probably made her immune against the post-syphilitic diseases, as is stated by some authors.

In conclusion I desire to express my thanks to Prof. Hugh T. Patrick, who so generously placed the material at my disposal, and who offered me most valuable suggestions in the preparation of this article. I also wish to thank Dr. Thos. A. Woodruff³² for his valuable ophthalmic records.

NOTE.—When the manuscript for this article was completed my attention was called to the fact that Dr. H. N. Moyer³³ has had an opportunity of examining the entire C. family in 1899. His report mentions distinct Argyll-Robertson pupils in the case of juvenile tabes and a positive diagnosis of general paresis in the case of T.—the Doctor having examined the boy during life. L., now the subject of hemiplegia and epilepsy, was perfectly well at that time.

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AN ADDITIONAL CASE OF PRECOCIOUS TABES.

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The following case, from the practice of Dr. Hugh T. Patrick, may be of some interest in connection with the preceding report. The case itself is rather unusual, as one of early tabes without evidence of syphilitic infection, with an exceptionally long course and with fragilitas ossium.

N. J. C., fifty years old, was seen September 3, 1900; family history negative. He is the oldest child, the next being seven years younger, and healthy. The patient has been married twenty-nine years, and has two healthy children of twenty and twenty-seven years respectively. Two children died—one at six weeks, of croup, the other quite young, of an accident.

The personal history is negative up to the beginning of his present trouble. When twenty-two years of age he began to suffer from sharp, shooting pains. They were severe and shifting, most severe in legs and arms, slighter and less frequent in the trunk and never invaded the head. At first these attacks occurred about once a month and lasted from a few hours to a day. Each one consisted of a sense of sharp pains, in duration from an instan-